

4:15 p.m.

831-2

Prevalence of Silent Pulmonary Emboli in Adults After the Fontan Operation

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Objectives. To determine the prevalence of pulmonary emboli (PE) in asymptomatic adult Fontan patients and identify the risk factors associated with PE.**Background.** Right atrial thrombi and systemic thromboembolic complications have been reported as a complication after the Fontan procedure. However the frequency of silent PE in this patient population is not known.**Methods.** All consecutive adult Fontan patients attending the adult congenital clinic over a six-month period underwent ventilation-perfusion (VQ) scanning and bloods for thrombophilia tendency. If the VQ scan showed an intermediate or high probability for PE, a spiral computerised tomography scan was performed to confirm the presence of PE.**Results.** Thirty patients (mean age 26 ± 7 y, 57% male) were included in this study. Five (17%) adult Fontan patients had an intermediate or high probability for PE on VQ scan, all of which were confirmed on spiral CT. No patient had a thrombophilia tendency. PE was not present in any patients (30%) taking warfarin. Late age at time of Fontan operation (19 ± 6 y vs. 11 ± 6 y, $p=0.012$) and lateral tunnel (3/4, $p=0.001$) were associated with increased risk of silent PE.**Conclusions.** Seventeen percent of adult patients with Fontan procedure have clinically silent PE. The hemodynamic long-term implications of this with respect to Fontan attrition over time are unknown. Large randomized prospective studies looking at anticoagulation therapy in all Fontan patients are urgently needed.

4:30 p.m.

831-3

Myocardial Blood Flow and Coronary Flow Reserve of the Anatomic Right Systemic Ventricle in Patients With Congenitally Corrected Transposition

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Objectives. Myocardial blood flow (MBF) in unoperated patients with congenitally corrected transposition of the great arteries (CCTGA) was investigated quantitatively by positron emission tomography (PET).**Background.** In CCTGA the systemic ventricle is of right morphology and patients are at high risk of developing systemic ventricular dysfunction; inadequate myocardial perfusion of the hypertrophied systemic ventricle may cause or accelerate a decline in ventricular function.**Methods.** Fifteen patients with CCTGA were investigated by PET with nitrogen-13 ammonia at rest and during adenosine vasodilatation. A subgroup of 7 patients had isolated CCTGA (group A: 30.3 ± 11.9 years), while the remaining 8 patients had complex CCTGA associated with subpulmonary stenosis in all, and ventricular septal defect in four (group B: 30.6 ± 16.4 years). Eleven healthy adults (26.2 ± 5.1 years) served as the control group.**Results.** Resting MBF was not different between both groups of patients with CCTGA and healthy young adults. Hyperemic blood flows were significantly lower in both groups of CCTGA compared to normals (195 ± 21 ml/100g/min. for group A, 201 ± 27 for group B, 258 ± 22 for normals; $p<0.001$); thus, coronary flow reserve was significantly lower in both groups of CCTGA compared to healthy adults (2.5 ± 0.28 for group A, 2.6 ± 0.48 for group B, 4.0 ± 0.43 for normals; $p<0.001$).**Conclusion.** Blood flow measurements suggest decreased coronary reserve in the absence of ischemic symptoms in patients with isolated and complex CCTGA. The global impairment of stress flow dynamics may indicate altered global vasoreactivity and quantitative changes in microcirculation suggest an important role in the pathogenesis of systemic RV dysfunction.

4:45 p.m.

831-4

Exercise-Related Neurohumoral Activation in Adults With Repaired Tetralogy of Fallot: A Pilot Study

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Background. Neurohumoral activation (NHA) in the setting of right ventricle (RV) dysfunction is poorly understood. We characterized resting and maximal voluntary exercise proANP (atrial natriuretic peptide), proBNP (brain natriuretic peptide) and ET I (endothelin) levels in adult patients with repaired Tetralogy of Fallot (TOF).**Patients & methods.** 20 consecutive patients >18 years, were prospectively evaluated with clinical assessment, 12-lead ECG, 2D-echo, MRI and graded bicycle cardiopulmonary exercise testing.**Results.** Median age was 30 years (range 20-48), and 10 were female. Median age at repair was 7.5 years (range 3-21).

	Resting Mean \pm SD (fmol/l)	Exercise Mean \pm SD (fmol/l)	p	Normal Range (fmol/l)	Rest vs. Exercise Abnormal counts %
proANP	2867 \pm 1930	3510 \pm 1805	0.001**	103-1359	68 vs. 79
proBNP	350 \pm 125	447 \pm 254	0.075**	126-366	47 vs. 68
endothelin I	1.5 \pm 1.3	1.5 \pm 1.4	0.58	0.12-0.7	47 vs. 47

Exercise induced proANP elevation correlated with longer cardiopulmonary bypass time ($r=0.46$, $p=0.004$), harder work of breathing ($r=0.35$, $p=0.04$), higher VO_2 peak ($r=0.40$, $p=0.016$) and greater exercise workload ($r=0.42$, $p=0.013$). Exercise related increases in proBNP correlated with higher VO_2 peak ($r=0.31$, $p=0.035$), and ECG markers of RVH i.e. greater R-wave height in V_1 ($r=0.35$, $p=0.04$). ET₁ levels at rest tended to correlate with greater exercise workload ($r=0.34$, $p=0.06$), and reduced work of breathing ($r=0.31$, $p=0.09$).**Conclusions.** NHA occurs at rest and during exercise in adult repaired TOF patients. It appears to be greatest in those patients with unfavorable peri-operative parameters, in those sustaining larger and longer exercise loads, and in those with bigger RV mass. NHA may be an important marker of resting and acute exercise induced RV hemodynamic stress.

5:00 p.m.

831-5

Outcome of Pulmonary Valve Replacement in Adults After Tetralogy Repair

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Background: There are a large number of adults with moderate/severe pulmonary regurgitation (PR) after tetralogy repair. An increasing number have had pulmonary valve replacement (PVR), but the results of this procedure have not been studied extensively. **Methods:** Accordingly the project committee of the International Society of Adult Congenital Heart Disease (ISACCD) undertook a retrospective analysis of the results of PVR. Seven centers participated in submitting data on 93 patients > 18 years of age who had PVR.**Results:** Average age at PVR was 26 ± 11 yr (median 27 yr) and median time of followup after PVR was 3 yr (range 4 days-28 yr). There were 2 deaths at 6 and 12 months after PVR. Right ventricular size estimated from echocardiography decreased in 81% ($p < 0.001$ testing for equal proportions) but RV systolic function, improvement in CHF status, increased in only 36% ($p=0.09$) from pre to postop study. Ability index improved in 59%, $p < 0.001$ and clinical CHF status improved in 22% ($p=0.01$). Kaplan-Meier estimates of durability of PVR showed approximately 50% replacement at 11 years.**Conclusion:** PVR is associated with a low mortality, a decrease in RV size, an improvement in ability index, an uncertain effect on RV systolic function, and an average valve durability of approximately 10 years. Criteria for PVR which will preserve RV function are not clearly identified and management of these patients remains a difficult enterprise.

5:15 p.m.

831-6

Right Ventricular Systolic Function Improvement After Pulmonary Valve Replacement in the Restricted Right Ventricle of Repaired Fallot Patients

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Background: Pulmonary regurgitation (PR) in patients late after total correction for Tetralogy of Fallot (TOF) may lead to progressive right ventricular (RV) systolic and diastolic dysfunction. Recently we assessed the beneficial effects of pulmonary valve replacement (PVR) on systolic RV function. Purpose of this study was to assess the influence of RV restriction on systolic function improvement following PVR, using MRI.**Methods:** Twentythree consecutive adult TOF patients who underwent PVR in our institution between 1998 and 2001 were studied. Median age at initial repair was 5.8 ± 4.5 years (range 0.4 to 21.0) and median age at PVR was 31.2 ± 9.4 years (range 17.0 to 45.6). Cardiac MRI was performed 6.1 ± 3.4 months before and 7.8 ± 2.2 and 20.3 ± 3.3 months after PVR. RV volumes were measured in the short axis and flow was measured through the pulmonary and tricuspid valve. RV restriction was defined as a significant (>5ml) end-diastolic forward flow (EDFF) in the main pulmonary artery.**Results:** Before PVR, 8 patients showed RV restriction. PR and RV end-diastolic volume (indexed for body surface area) EDVI were not different in the restriction group compared to the non-restriction group, respectively 46 ± 10 ml vs. 45 ± 11 ml and 171 ± 26 ml/m² vs. 161 ± 44 ml/m². Twenty months after PVR, 9 patients had residual PR (mean 12 ± 7 ml); five of them had EDFF before PVR. Of the 14 patients without residual PR, only 3 had EDFF before PVR ($P<0.1$). The presence of residual PR was associated with less total decrease of RV EDVI at 20 months, respectively $40 \pm 10\%$ and $30 \pm 10\%$, while at 8 months post-PVR, there was no significant difference of improvement; respectively $35 \pm 12\%$ and $32 \pm 11\%$.**Conclusion:** In adult patients late after total correction of TOF, restriction of the RV tends to be associated with an increased chance of residual PR after surgery. Residual PR after PVR prohibits further improvement of RV EDVI after initial improvement.